#2

```
            RAW SEQUENCE LISTING                    DATE: 07/23/2001
            PATENT APPLICATION:  US/09/895,072      TIME: 13:19:44

            Input Set : A:\210119USOCONT.txt
            Output Set: N:\CRF3\07232001\I895072.raw
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ENTERED
See page 5

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        3 <110> APPLICANT: CANFIELD, WILLIAM M
        5 <120> TITLE OF INVENTION: METHODS FOR PRODUCING HIGHLY PHOSPHORYLATED LYSOSOMAL
HYDROLASES
        7 <130> FILE REFERENCE: 210119USOCONT
C-->    9 <140> CURRENT APPLICATION NUMBER: US/09/895,072
C-->    9 <141> CURRENT FILING DATE: 2001-07-02
        9 <150> PRIOR APPLICATION NUMBER: 60/153,831
       10 <151> PRIOR FILING DATE: 1999-09-14
       12 <150> PRIOR APPLICATION NUMBER: US 09/635,872
       13 <151> PRIOR FILING DATE: 2000-08-10
       15 <160> NUMBER OF SEQ ID NOS: 52
       17 <170> SOFTWARE: PatentIn version 3.1
       19 <210> SEQ ID NO: 1
       20 <211> LENGTH: 928
       21 <212> TYPE: PRT
       22 <213> ORGANISM: Homo sapiens
       24 <400> SEQUENCE: 1
       26 Met Leu Phe Lys Leu Leu Gln Arg Gln Thr Tyr Thr Cys Leu Ser His
       27 1               5                   10                  15
       30 Arg Tyr Gly Leu Tyr Val Cys Phe Leu Gly Val Val Val Thr Ile Val
       31             20                  25                  30
       34 Ser Ala Phe Gln Phe Gly Glu Val Val Leu Glu Trp Ser Arg Asp Gln
       35         35                  40                  45
       38 Tyr His Val Leu Phe Asp Ser Tyr Arg Asp Asn Ile Ala Gly Lys Ser
       39     50                  55                  60
       42 Phe Gln Asn Arg Leu Cys Leu Pro Met Pro Ile Asp Val Val Tyr Thr
       43 65                  70                  75                  80
       46 Trp Val Asn Gly Thr Asp Leu Glu Leu Leu Lys Glu Leu Gln Gln Val
       47             85                  90                  95
       50 Arg Glu Gln Met Glu Glu Glu Gln Lys Ala Met Arg Glu Ile Leu Gly
       51             100                 105                 110
       54 Lys Asn Thr Thr Glu Pro Thr Lys Lys Ser Glu Lys Gln Leu Glu Cys
       55         115                 120                 125
       58 Leu Leu Thr His Cys Ile Lys Val Pro Met Leu Val Leu Asp Pro Ala
       59     130                 135                 140
       62 Leu Pro Ala Asn Ile Thr Leu Lys Asp Val Pro Ser Leu Tyr Pro Ser
       63 145                 150                 155                 160
       66 Phe His Ser Ala Ser Asp Ile Phe Asn Val Ala Lys Pro Lys Asn Pro
       67             165                 170                 175
       70 Ser Thr Asn Val Ser Val Val Val Phe Asp Ser Thr Lys Asp Val Glu
       71             180                 185                 190
       74 Asp Ala His Ser Gly Leu Leu Lys Gly Asn Ser Arg Gln Thr Val Trp
       75         195                 200                 205
       78 Arg Gly Tyr Leu Thr Thr Asp Lys Glu Val Pro Gly Leu Val Leu Met
       79     210                 215                 220
       82 Gln Asp Leu Ala Phe Leu Ser Gly Phe Pro Pro Thr Phe Lys Glu Thr
       83 225                 230                 235                 240
       86 Asn Gln Leu Lys Thr Lys Leu Pro Glu Asn Leu Ser Ser Lys Val Lys
```

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 87                 245                250                255
 90 Leu Leu Gln Leu Tyr Ser Glu Ala Ser Val Ala Leu Leu Lys Leu Asn
 91             260                265                270
 94 Asn Pro Lys Asp Phe Gln Glu Leu Asn Lys Gln Thr Lys Lys Asn Met
 95         275                280                285
 98 Thr Ile Asp Gly Lys Glu Leu Thr Ile Ser Pro Ala Tyr Leu Leu Trp
 99     290                295                300
102 Asp Leu Ser Ala Ile Ser Gln Ser Lys Gln Asp Glu Asp Ile Ser Ala
103 305                310                315                320
106 Ser Arg Phe Glu Asp Asn Glu Glu Leu Arg Tyr Ser Leu Arg Ser Ile
107             325                330                335
110 Glu Arg His Ala Pro Trp Val Arg Asn Ile Phe Ile Val Thr Asn Gly
111         340                345                350
114 Gln Ile Pro Ser Trp Leu Asn Leu Asp Asn Pro Arg Val Thr Ile Val
115         355                360                365
118 Thr His Gln Asp Val Phe Arg Asn Leu Ser His Leu Pro Thr Phe Ser
119     370                375                380
122 Ser Pro Ala Ile Glu Ser His Ile His Arg Ile Glu Gly Leu Ser Gln
123 385                390                395                400
126 Lys Phe Ile Tyr Leu Asn Asp Asp Val Met Phe Gly Lys Asp Val Trp
127             405                410                415
130 Pro Asp Asp Phe Tyr Ser His Ser Lys Gly Gln Lys Val Tyr Leu Thr
131         420                425                430
134 Trp Pro Val Pro Asn Cys Ala Glu Gly Cys Pro Gly Ser Trp Ile Lys
135         435                440                445
138 Asp Gly Tyr Cys Asp Lys Ala Cys Asn Asn Ser Ala Cys Asp Trp Asp
139     450                455                460
142 Gly Gly Asp Cys Ser Gly Asn Ser Gly Gly Ser Arg Tyr Ile Ala Gly
143 465                470                475                480
146 Gly Gly Gly Thr Gly Ser Ile Gly Val Gly His Pro Trp Gln Phe Gly
147             485                490                495
150 Gly Gly Ile Asn Ser Val Ser Tyr Cys Asn Gln Gly Cys Ala Asn Ser
151         500                505                510
154 Trp Leu Ala Asp Lys Phe Cys Asp Gln Ala Cys Asn Val Leu Ser Cys
155         515                520                525
158 Gly Phe Asp Ala Gly Asp Cys Gly Gln Asp His Phe His Glu Leu Tyr
159     530                535                540
162 Lys Val Ile Leu Leu Pro Asn Gln Thr His Tyr Ile Ile Pro Lys Gly
163 545                550                555                560
166 Glu Cys Leu Pro Tyr Phe Ser Phe Ala Glu Val Ala Lys Arg Gly Val
167         565                570                575
170 Glu Gly Ala Tyr Ser Asp Asn Pro Ile Ile Arg His Ala Ser Ile Ala
171     580                585                590
174 Asn Lys Trp Lys Thr Ile His Leu Ile Met His Ser Gly Met Asn Ala
175     595                600                605
178 Thr Thr Ile His Phe Asn Leu Thr Phe Gln Asn Thr Asn Asp Glu Glu
179     610                615                620
182 Phe Lys Met Gln Ile Thr Val Glu Val Asp Thr Arg Glu Gly Pro Lys
183 625                630                635                640
```

Input Set : A:\210119US0CONT.txt
Output Set: N:\CRF3\07232001\I895072.raw

```
186 Leu Asn Ser Thr Ala Gln Lys Gly Tyr Glu Asn Leu Val Ser Pro Ile
187             645                 650                 655
190 Thr Leu Leu Pro Glu Ala Glu Ile Leu Phe Glu Asp Ile Pro Lys Glu
191             660                 665                 670
194 Lys Arg Phe Pro Lys Phe Lys Arg His Asp Val Asn Ser Thr Arg Arg
195         675                 680                 685
198 Ala Gln Glu Glu Val Lys Ile Pro Leu Val Asn Ile Ser Leu Leu Pro
199     690                 695                 700
202 Lys Asp Ala Gln Leu Ser Leu Asn Thr Leu Asp Leu Gln Leu Glu His
203 705                 710                 715                 720
206 Gly Asp Ile Thr Leu Lys Gly Tyr Asn Leu Ser Lys Ser Ala Leu Leu
207             725                 730                 735
210 Arg Ser Phe Leu Met Asn Ser Gln His Ala Lys Ile Lys Asn Gln Ala
211             740                 745                 750
214 Ile Ile Thr Asp Glu Thr Asn Asp Ser Leu Val Ala Pro Gln Glu Lys
215             755                 760                 765
218 Gln Val His Lys Ser Ile Leu Pro Asn Ser Leu Gly Val Ser Glu Arg
219     770                 775                 780
222 Leu Gln Arg Leu Thr Phe Pro Ala Val Ser Val Lys Val Asn Gly His
223 785                 790                 795                 800
226 Asp Gln Gly Gln Asn Pro Pro Leu Asp Leu Glu Thr Thr Ala Arg Phe
227             805                 810                 815
230 Arg Val Glu Thr His Thr Gln Lys Thr Ile Gly Gly Asn Val Thr Lys
231             820                 825                 830
234 Glu Lys Pro Pro Ser Leu Ile Val Pro Leu Glu Ser Gln Met Thr Lys
235             835                 840                 845
238 Glu Lys Lys Ile Thr Gly Lys Glu Lys Glu Asn Ser Arg Met Glu Glu
239     850                 855                 860
242 Asn Ala Glu Asn His Ile Gly Val Thr Glu Val Leu Leu Gly Arg Lys
243 865                 870                 875                 880
246 Leu Gln His Tyr Thr Asp Ser Tyr Leu Gly Phe Leu Pro Trp Glu Lys
247             885                 890                 895
250 Lys Lys Tyr Phe Gln Asp Leu Leu Asp Glu Glu Glu Ser Leu Lys Thr
251         900                 905                 910
254 Gln Leu Ala Tyr Phe Thr Asp Ser Lys Asn Thr Gly Arg Gln Leu Lys
255         915                 920                 925
258 <210> SEQ ID NO: 2
259 <211> LENGTH: 328
260 <212> TYPE: PRT
261 <213> ORGANISM: Homo sapiens
263 <400> SEQUENCE: 2
265 Asp Thr Phe Ala Asp Ser Leu Arg Tyr Val Asn Lys Ile Leu Asn Ser
266 1               5                   10                  15
269 Lys Phe Gly Phe Thr Ser Arg Lys Val Pro Ala His Met Pro His Met
270             20                  25                  30
273 Ile Asp Arg Ile Val Met Gln Glu Leu Gln Asp Met Phe Pro Glu Glu
274         35                  40                  45
277 Phe Asp Lys Thr Ser Phe His Lys Val Arg His Ser Glu Asp Met Gln
278     50                  55                  60
```

RAW SEQUENCE LISTING   DATE: 07/23/2001
PATENT APPLICATION: US/09/895,072   TIME: 13:19:44

Input Set : A:\210119US0CONT.txt
Output Set: N:\CRF3\07232001\I895072.raw

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281 Phe Ala Phe Ser Tyr Phe Tyr Tyr Leu Met Ser Ala Val Gln Pro Leu
282 65               70                  75                  80
285 Asn Ile Ser Gln Val Phe Asp Glu Val Asp Thr Asp Gln Ser Gly Val
286                  85                  90                  95
289 Leu Ser Asp Arg Glu Ile Arg Thr Leu Ala Thr Arg Ile His Glu Leu
290              100                 105                 110
293 Pro Leu Ser Leu Gln Asp Leu Thr Gly Leu Glu His Met Leu Ile Asn
294              115                 120                 125
297 Cys Ser Lys Met Leu Pro Ala Asp Ile Thr Gln Leu Asn Asn Ile Pro
298          130              135                 140
301 Pro Thr Gln Glu Ser Tyr Tyr Asp Pro Asn Leu Pro Pro Val Thr Lys
302 145                  150                 155                 160
305 Ser Leu Val Thr Asn Cys Lys Pro Val Thr Asp Lys Ile His Lys Ala
306              165                 170                 175
309 Tyr Lys Asp Lys Asn Lys Tyr Arg Phe Glu Ile Met Gly Glu Glu Glu
310              180                 185                 190
313 Ile Ala Phe Lys Met Ile Arg Thr Asn Val Ser His Val Val Gly Gln
314          195                 200                 205
317 Leu Asp Asp Ile Arg Lys Asn Pro Arg Lys Phe Val Cys Leu Asn Asp
318          210                 215                 220
321 Asn Ile Asp His Asn His Lys Asp Ala Gln Thr Val Lys Ala Val Leu
322 225                  230                 235                 240
325 Arg Asp Phe Tyr Glu Ser Met Phe Pro Ile Pro Ser Gln Phe Glu Leu
326              245                 250                 255
329 Pro Arg Glu Tyr Arg Asn Arg Phe Leu His Met His Glu Leu Gln Glu
330              260                 265                 270
333 Trp Arg Ala Tyr Arg Asp Lys Leu Lys Phe Trp Thr His Cys Val Leu
334          275                 280                 285
337 Ala Thr Leu Ile Met Phe Thr Ile Phe Ser Phe Phe Ala Glu Gln Leu
338      290                 295                 300
341 Ile Ala Leu Lys Arg Lys Ile Phe Pro Arg Arg Arg Ile His Lys Glu
342 305                 310                 315                 320
345 Ala Ser Pro Asn Arg Ile Arg Val
346                  325
```

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349 <210> SEQ ID NO: 3
350 <211> LENGTH: 305
351 <212> TYPE: PRT
352 <213> ORGANISM: Homo sapiens
354 <220> FEATURE:
355 <221> NAME/KEY: SIGNAL
356 <222> LOCATION: (1)..(24)
357 <223> OTHER INFORMATION:
360 <400> SEQUENCE: 3
362 Met Ala Ala Gly Leu Ala Arg Leu Leu Leu Leu Leu Gly Leu Ser Ala
363 1               5                   10                  15
366 Gly Gly Pro Ala Pro Ala Gly Ala Ala Lys Met Lys Val Val Glu Glu
367              20                  25                  30
370 Pro Asn Ala Phe Gly Val Asn Asn Pro Phe Leu Pro Gln Ala Ser Arg
371          35                  40                  45
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RAW SEQUENCE LISTING   DATE: 07/23/2001
PATENT APPLICATION: US/09/895,072   TIME: 13:19:44

Input Set : A:\210119US0CONT.txt
Output Set: N:\CRF3\07232001\I895072.raw

```
374 Leu Gln Ala Lys Arg Asp Pro Ser Pro Val Ser Gly Pro Val His Leu
375      50              55              60
378 Phe Arg Leu Ser Gly Lys Cys Phe Ser Leu Val Glu Ser Thr Tyr Lys
379 65              70              75                  80
382 Tyr Glu Phe Cys Pro Phe His Asn Val Thr Gln His Glu Gln Thr Phe
383              85              90              95
386 Arg Trp Asn Ala Tyr Ser Gly Ile Leu Gly Ile Trp His Glu Trp Glu
387          100              105              110
390 Ile Ala Asn Asn Thr Phe Thr Gly Met Trp Met Arg Asp Gly Asp Ala
391          115              120              125
394 Cys Arg Ser Arg Ser Arg Gln Ser Lys Val Glu Leu Ala Cys Gly Lys
395      130              135              140
398 Ser Asn Arg Leu Ala His Val Ser Glu Pro Ser Thr Cys Val Tyr Ala
399 145              150              155                  160
402 Leu Thr Phe Glu Thr Pro Leu Val Cys His Pro His Ala Leu Leu Val
403              165              170              175
406 Tyr Pro Thr Leu Pro Glu Ala Leu Gln Arg Gln Trp Asp Gln Val Glu
407              180              185              190
410 Gln Asp Leu Ala Asp Glu Leu Ile Thr Pro Gln Gly His Glu Lys Leu
411          195              200              205
414 Leu Arg Thr Leu Phe Glu Asp Ala Gly Tyr Leu Lys Thr Pro Glu Glu
415      210              215              220
418 Asn Glu Pro Thr Gln Leu Glu Gly Gly Pro Asp Ser Leu Gly Phe Glu
419 225              230              235                  240
422 Thr Leu Glu Asn Cys Arg Lys Ala His Lys Glu Leu Ser Lys Glu Ile
423              245              250              255
426 Lys Arg Leu Lys Gly Leu Leu Thr Gln His Gly Ile Pro Tyr Thr Arg
427          260              265              270
430 Pro Thr Glu Thr Ser Asn Leu Glu His Leu Gly His Glu Thr Pro Arg
431          275              280              285
434 Ala Lys Ser Pro Glu Gln Leu Arg Gly Asp Pro Gly Leu Arg Gly Ser
435      290              295              300
438 Leu
439 305
442 <210> SEQ ID NO: 4
443 <211> LENGTH: 5597
444 <212> TYPE: DNA
445 <213> ORGANISM: Homo sapiens
447 <400> SEQUENCE: 4
448 cggagccgag cgggcgtccg tcgccggagc tgcaatgagc ggcgcccgga ggctgtgacc      60
450 tgcgcgcggc ggcccgaccg gggcccctga atggcggctc gctgaggcgg cggcggcggc     120
452 ggcggctcag gctcctcggg gcgtggcgtg gcggtgaagg ggtgatgctg ttcaagctcc     180
454 tgcagagaca aacctatacc tgcctgtccc acaggtatgg gctctacgtg tgcttcttgg     240
456 gcgtcgttgt caccatcgtc tccgccttcc agttcggaga ggtggttctg gaatggagcc     300
458 gagatcaata ccatgttttg tttgattcct atagagacaa tattgctgga aagtcctttc     360
460 agaatcggct ttgtctgccc atgccgattg acgttgttta cacctgggtg aatggcacag     420
462 atcttgaact actgaaggaa ctacagcagg tcagagaaca gatggaggag gagcagaaag     480
464 caatgagaga aatccttggg aaaaacacaa cggaacctac taagaagagt gagaagcagt     540
466 tagagtgttt gctaacacac tgcattaagg tgccaatgct tgtactggac ccagccctgc     600
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VERIFICATION SUMMARY   DATE: 07/23/2001
PATENT APPLICATION: US/09/895,072   TIME: 13:19:45

Input Set : **A:\210119US0CONT.txt**
Output Set: **N:\CRF3\07232001\I895072.raw**

```
L:9 M:270 C: Current Application Number differs, Replaced Current Application No
L:9 M:271 C: Current Filing Date differs, Replaced Current Filing Date
L:1120 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:10
L:2207 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:17
L:2211 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:17
L:2213 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:17
L:2754 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:23
L:2926 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:35
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